

Muco-submucosal elongated polyps of the gastrointestinal tract: A case series and a review of the literature

Char Loo Tan, Sze Hwa Tan, Jimmy BY So, Fredrik Petersson

Char Loo Tan, Sze Hwa Tan, Fredrik Petersson, Department of Pathology, National University Hospital System, Singapore 119074, Singapore

Jimmy BY So, Department of Surgery, National University Hospital System, Singapore 119074, Singapore

Author contributions: Tan CL, Tan SH, So JBY, Petersson F contributed to the manuscript writing and revision.

Correspondence to: Fredrik Petersson, MD, PhD, Department of Pathology, National University Health System, 5 Lower Kent Ridge Road, Singapore 119074, Singapore. fredrikpetersson@live.se

Telephone: +65-97-714890 Fax: +65-67-780671

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Abstract

We present three cases of gastrointestinal muco-submucosal elongated polyps, two located in the duodenum and one in the descending colon. All three cases had a characteristic, "worm-like" endoscopic appearance and were lined by unremarkable mucosa. The vascular component was located in the submucosa and was composed of a mixture of variably dilated blood vessels (capillaries and veins) and lymphatics. The duodenal polyps displayed lipomatous metaplasia of the submucosal stroma. The dual vascular phenotype of the vascular component was confirmed by immunohistochemistry with D2-40 and CD31.

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Key words: Muco-submucosal elongated polyp; Vascular malformation; Endoscopic ultrasonography; Angioma; Endoscopy

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INTRODUCTION

Benign gastrointestinal polyps composed of mucosa and submucosa, the latter with a prominent vascular component are relatively unusual non-neoplastic lesions with uncertain etiopathogenesis. Such polyps have mainly been studied by Japanese investigators, but are increasingly being recognized by western investigators^[1,2]. The nomenclature pertaining to these polyps has varied. In the seminal paper on this entity, Mataka *et al*^[3] chose the term "colonic muco-submucosal elongated polyp". Most of the reported cases have been located in the large bowel^[4-7], but three polyps with identical endoscopic and histological features have also been documented in the small bowel, including the duodenum^[8-10].

Irrespective of site, muco-submucosal elongated polyps display characteristic endoscopic features, with a "worm-like" appearance which are lined by unremarkable mucosa. Histologic examination confirms the presence of a normal mucosa and reveals a submucosal component with a variably prominent mixture of blood vessels and lymphatics and absence of significant inflammation. In this paper we present the clinicopathologic features of three additional cases of this entity and review the literature on these characteristic benign gastrointestinal polyps.

CASE REPORT

Case 1

A 55 year-old previously healthy female presented with postprandial abdominal discomfort of 3 mo duration. There were no signs or symptoms of gastrointestinal bleeding or malabsorption. Gastroduodenoscopy revealed a 4 cm long, slender, "worm-like" polyp in the second part of the duodenum (Figure 1A). The polyp was

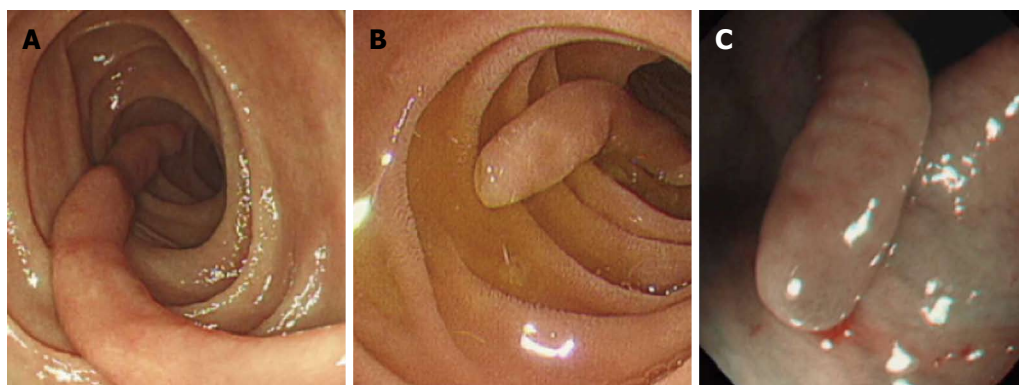


Figure 1 Endoscopic views of the polyps. A: Gastroduodenoscopy revealed a 4 cm long, slender, “worm-like” polyp in the second part of the duodenum; B: Gastroduodenoscopy showed a 2.2 cm long, slender polyp in the duodenum; C: Colonoscopy showed a 1.4 cm long, slender polyp in the descending colon.

removed endoscopically. The patient was well on follow up at 3 mo.

Case 2

A 70 year-old previously healthy man presented with reflux symptoms of unknown duration. Gastroduodenoscopy showed a 2.2 cm long, slender polyp in the duodenum (Figure 1B). The polyp was removed endoscopically.

Case 3

A 74 year-old female with a history of hypertension and iron deficiency anemia secondary to peptic ulcer disease presented with epigastric pain associated with constipation of 1 mo duration. Colonoscopy showed a 1.4 cm long, slender polyp in the descending colon (Figure 1C) and a distal rectal ulcer near the anal verge. The polyp was removed endoscopically. There was no evidence of diverticular disease. In addition, gastroduodenoscopy showed several benign gastric and duodenal ulcers (biopsies from the gastric ulcers showed mild chronic gastritis with reactive changes and no activity; no *Helicobacter pylori* were identified; the rectal biopsy showed features consistent with a solitary rectal ulcer).

The tissues were fixed in neutral formalin and both polyps were completely embedded in paraffin. 4 µm thick sections were cut and stained with hematoxylin and eosin (HE). An immunohistochemical study with commercially available antibodies (D2-40-podoplanin, CD31) using protocols according to the manufacturers’ recommendations were employed in Case 1 and 3. Immunohistochemistry with WT-1 were performed in all three cases.

Gross findings and histology

All three polyps were thin and elongated corresponding to the “worm-like” endoscopic appearance (Figure 2A). All polyps were lined by unremarkable mucosa. The submucosal components contained prominent vasculature including dilated, variably sized veins and lymphatic vessels running parallel to the long axis of the polyps and surrounded by loose collagenous stroma (Figure 2B and C). In addition, there was also focal lipomatous metaplasia in both duodenal polyps. No arterial vascular com-

ponent was identified. No vascular abnormalities were present in the mucosa. No significant inflammation was identified.

Immunohistochemistry

The endothelial cells of both vascular components in the polyps displayed immunoreactivity for CD31 throughout the lesion (Figure 3A). D2-40-podoplanin was selectively expressed by the endothelial cells of the lymphatic component (Figure 3B). In Case 1 and 2, the endothelial cells of both vascular components displayed patchy cytoplasmic positivity for WT-1 whereas Case 3 was completely negative (data not shown).

DISCUSSION

We herein report three cases of muco-submucosal elongated polyps, two in the duodenum and the other one in descending colon. This characteristic, albeit unusual entity, is not well covered in most standard textbooks in gastrointestinal pathology. Initially described by Japanese investigators, Matake *et al.*^{3,11} published two series of these polyps comprising 19 cases *in toto*. The polyps ranged in size from 1.2-16 cm (mean 2.9 cm) and displayed a characteristic appearance (elongated, slender, drumstick shaped). Histological examination revealed normal mucosa and a loose to dense submucosal layer containing a variably prominent vascular component composed of dilated blood vessels (veins and capillaries) and lymphatics. Subsequently, these polyps have been increasingly recognized and mostly documented in the large bowel (all parts) including rectum^{1,5,6,12,13}. In all 23 reported polyps from the large bowel, the size has ranged from 0.7 to 15 cm (mean 2.8 cm).

Two polyps with endoscopic and morphologic features conforming to a muco-submucosal elongated polyp have also been described in the small bowel^{9,10}.

In addition, in 2009, Kim *et al.*⁸ reported a case of a 5 cm long duodenal polyp with a “wormlike/drumstick” endoscopic appearance which they labelled as “polypoid vascular and lymphatic malformation of the duodenum”. The histologic features as described by the authors and the provided photomicrographs show, in our opinion, a

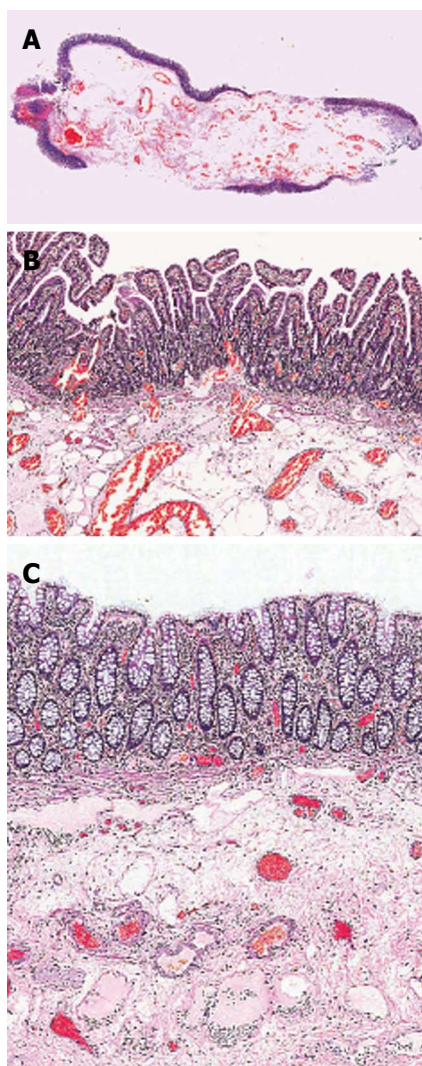


Figure 2 Histological section (hematoxylin and eosin staining). A: Case 1; B: Case 1 with normal small intestinal mucosal lining; C: Case 3 with normal large bowel mucosa overlying the submucosa which contains a prominent vascular component.

muco-submucosal elongated polyp.

Our Case 1 and 2 were of duodenal origin, which is a rare site and thus represents the third and fourth published cases of muco-submucosal elongated polyps of the small bowel.

A lesion resembling muco-submucosal elongated polyps has been described by Kelly^[14]. In this paper, the author characterized eight cases of swollen mucosal folds and broad based, ‘leaf-like’ polyps with mucosal and submucosal vascular congestion, thrombi, edema, hemorrhage and hemosiderin deposits in the sigmoid colon associated with diverticular disease in resection specimens. In addition to the submucosal component, these polypoidal lesions showed mucosal prolapse-type changes. The author adequately labelled these lesions “polypoid prolapsing mucosal folds in diverticular disease” (PPMF). Interestingly, cases of muco-submucosal elongated polyps associated with diverticular disease have also been documented^[1,7]. In contrast to muco-submucosal

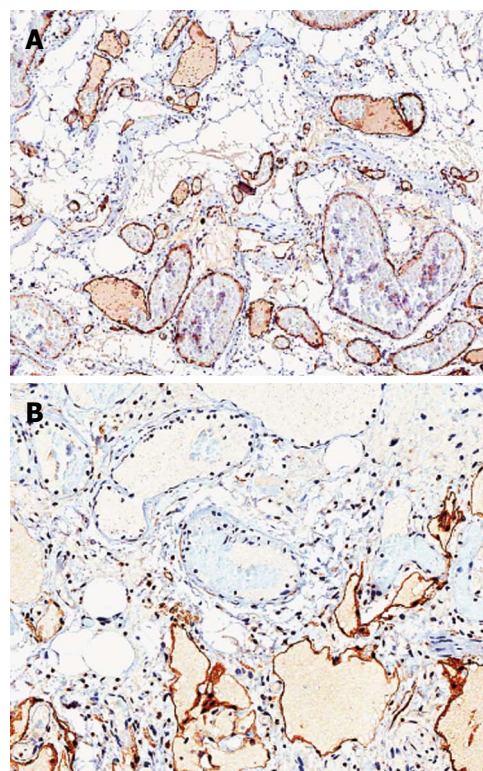


Figure 3 Immunohistochemistry. A: Positivity for CD31 in the endothelial cells of both vascular components; B: Positivity for D2-40/podoplanin selectively expressed by the endothelial cells in the lymphatic vessels.

elongated polyps, PPMFs do not display the “wormlike”, slender gross appearance with narrow attachment to the bowel mucosa. In addition, on histological examination, the mucosal component in PPMFs shows prolapse-type changes, which reportedly are absent in muco-submucosal elongated polyps, including the three cases presented herein.

Absence of significant inflammation in muco-submucosal elongated polyps also differentiate this entity from other mucosal prolapse associated polyps such as inflammatory “cap” polyps, inflammatory cloacogenic polyps and inflammatory myoglandular polyps^[15].

Filiform polyps (a subtype of inflammatory polyps) characterized by finger like projection covered by normal colonic mucosa with central core containing vessels and smooth muscle fibers can also mimic muco-submucosal elongated polyps. However, these polyps are usually multiple (“filiform polyposis”) and are associated with inflammatory bowel disease. Of note is that cases without apparent history of inflammatory bowel disease appear to exist^[16].

An inverted colonic diverticulum is a clinically important differential diagnosis. When these are large in size, they may resemble a pedunculated polyp and an endoscopic polypectomy may be performed which is associated with risk of perforation^[17]. A high index of clinical suspicion is important for these lesions and the key to its recognition is the endoscopic association with conventional diverticuli in the segment where the polyp

is located. A small biopsy from an inverted diverticulum shows non-specific features including normal to mildly inflamed mucosa and congested submucosa with variable degrees of chronic inflammation.

The residual stalk after a previously excised pedunculated adenoma or after autoamputation of any pedunculated polyp may also come into the endoscopic and histological differential diagnosis. Distinguishing, albeit non-specific features, are procedure related changes (granulation tissue and hemosiderin) and obviously, a history of previous polypectomy.

The etiology of muco-submucosal elongated polyps is unknown. It has been suggested that the gastrointestinal peristaltic movement may serve as mechanical traction for redundant mucosa, thereby making such areas of mucosa a nidus for polyp formation^[3,18]. In line with this, Alizart *et al*^[11] hypothesized that focal areas with prominent submucosal venous plexus may elevate the mucosa and serve as the leading point/nidus for the traction and hence polyp formation. A similar explanation has also been put forward by Kelly^[14] to explain the formation of PPMFs associated with diverticular disease.

We have immunohistochemically confirmed that the submucosal vascular component is composed of both blood vessels and lymphatics. Thus, we suggest that in addition to the criteria for the diagnosis of muco-submucosal elongated polyp as suggested by Alizart *et al*^[11]: (1) elongated, cylindrical shape with narrow base; (2) presence of submucosa; (3) absence of marked architectural disturbance of the overlying mucosa; and (4) absence of inflammatory infiltrate in both the mucosa and submucosa; the presence of a significant vascular submucosal component with a dual phenotype could be added to these criteria.

The endoscopic ultrasonographic features of muco-submucosal elongated polyps have been reported by Takahashi *et al*^[12] and Akahoshi *et al*^[13]. The ultrasonographic findings appear characteristic for this entity and in all lesions, elongated polypoid structures with both mucosal and submucosal layers were visualized. The “microcystic components” that the authors reported in the submucosal layer, most likely corresponds to the ectatic vascular component that is characteristically present in these lesions. The authors conclude that the distinctive ultrasonographic features allow for the diagnosis to be made and hence differentiate muco-submucosal elongated polyps from other submucosal lesions such as leiomyoma, lipoma and lymphangioma^[13]. Successful endoscopic removal of muco-submucosal elongated polyp (as in our cases) have previously been reported^[5,6,9,13].

Recently it has been suggested that endothelial cytoplasmic immunohistochemical expression of WT-1 is helpful in discriminating between cutaneous vascular neoplasms and vascular malformations. Trindade *et al*^[19] studied 117 cutaneous vascular neoplasms and 50 vascular malformations and found that all vascular neoplasms showed diffuse endothelial expression of WT-1 whereas all vascular malformations except for the arteriovenous

type were negative. The expression of WT-1 in a random manner in our first and second case and complete negativity in our third case lend support to the reactive nature of the vascular components in muco-submucosal elongated polyps. However, it is important to bear in mind that all the cases in Trindade *et al*^[19] study were of cutaneous origin and the underlying molecular mechanisms behind the development of cutaneous and intestinal vascular lesions may differ. Further studies are warranted to clarify this.

In conclusion, we report three cases of gastrointestinal muco-submucosal elongated polyps with prominent submucosal vascular components. One of these was located in the large bowel and the other two were of duodenal origin. We have determined by immunohistochemistry that the vascular component is a mixture of blood vessels and lymphatics. The endoscopic ultrasonographic appearance of these polyps appears to be characteristic allowing for a high degree of certainty in making this diagnosis by the endoscopists.

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